ADAMANTINOMAS OF THE HYPOPHYSEAL STALK AND SPHENOID BONE

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Tumors of the hypophyseal stalk, of which the adamantinomas form the largest single group, are sufficiently rare to remain a pathological curiosity. Erdheim (1) in 1904 first made an extensive study and review of the subject and since then valuable contributions have been added by Duffy (2), Critchley and Ironside (3), and others.

Adamantinomas have been defined by Critchley and Ironside as a variety of epithelial tumors in the suprapituitary region arising from an unobliterated portion of the fetal craniopharyngeal duct. The histological structure resembles that of the embryonic enamel organ and bears a striking resemblance to the epithelial odontomas of the jaw.

REPORT OF CASES

CASE 1: History: L. L., aged three years, white female, had been well except for a history of whooping cough which was followed by pneumonia at the age of three months. Her birth history and development were normal except that she was exceptionally bright. She “understood and talked like a ten-year-old child.”

Three months prior to her entrance to the Research and Educational Hospitals of the University of Illinois, on Dec. 18, 1933, the patient suddenly started to vomit and exhibited a complete change of personality with peculiar actions, as grinding of the teeth, biting of the finger nails, pacing the floor, and at times singing, yelling or crying and talking irrationally. Her vision had been rapidly decreasing; the gait became tottering, and the head grew noticeably larger. Subsequently she became incontinent. She did not complain of headaches or dizziness at any time, but mentally grew progressively worse and ultimately reverted to an infantile state.

Examination revealed a well developed and well nourished child who did not appear acutely ill. The heart, lungs, and abdominal organs were essentially normal. The head was moderately hydrocephalic, and a positive Macawen’s sign (cracked-pot resonance) was

1 Read before the Chicago Pathological Society, Dec. 10, 1934.
obtained. There was almost total blindness. The pupils reacted sluggishly to light. The eyes were in midline with no apparent nystagmus. External ocular movements could not be definitely ascertained because of poor cooperation. Examination of the fundi revealed advanced bilateral optic atrophy with a superimposed choked disk. A slight left-side facial asymmetry was noticeable when the child cried. The tongue, when protruded, deviated to the left. A left hemiparesis was present with all the deep reflexes marked. Pathological reflexes were absent. Sensibility was intact throughout.

Roentgenograms revealed a suprasellar calcification with marked enlargement of the sella turcica, separation of the cranial sutures, and a mild hydrocephalus.

 Diagnosis: A suprasellar cyst, probably a craniopharyngioma, was suspected. A dermoid cyst or cholesteatoma was also considered.

![Figure 1](https://example.com/figure1.jpg)

**FIG. 1. CASE I: PHOTOMICROGRAPH SHOWING TOOTHBUD-LIKE OUTPOUCHING FROM THE EPITHELIAL CELL LAYERS**

B. Outer layer of cylindrical epithelial cells arranged in a parallel row. C. Calcification at the periphery. A. Arachnoid cell layer. Heidenhain's iron hematoxylin stain. × 15.

*Operation (Dr. Eric Oldberg)*: A right transfrontal bone flap was turned down and, without opening the dura, a ventricular needle was inserted into the right frontal lobe. At a depth of 1 cm. a huge cyst was encountered, containing a chocolate-colored fluid loaded with cholesterin crystals. On exploration, 300 c.c. of fluid was obtained, after which the flap was closed. The hypothalamic region was not attacked at that session.

*Postoperative Course*: Shortly after the operation a temperature of 104° F. developed. This gradually receded and the child improved. She sang and talked constantly, using the most obscene language. The left lower facial paresis disappeared, but the swelling of the disks had not receded. On the eleventh postoperative day scarlet fever developed and the patient was transferred to the Cook County Contagious Hospital, where she died after the occurrence of left-sided jacksonian convulsions.

*Necropsy (Dr. R. H. Jaffé)*: Externally there was a 7 × 6.5 cm. defect on the right frontal bone covered by a bone flap. Beneath the bone flap the dura mater was covered by a thin, yellow-brown to purplish-red membrane. The inside of the dura mater over
both hemispheres was covered by a thin, spiderweb-like, yellow-gray membrane which stretched across the subdural space to the leptomeninges over both cerebral hemispheres. On removal of the brain, 250 c.c. of a brownish and slightly clouded fluid escaped. The brain weighed 1220 gm. The region of the infundibulum was the site of an irregular, lobular, firm mass 17 mm. in transverse, 18 mm. in antero-posterior diameter, and 20 mm. in height. The mass consisted of light yellow-brown, irregular lobules and bordered anteriorly on the optic chiasm. It fitted into a groove in the body of the sphenoid bone which measured 20 × 17 × 7 mm. in diameter. The brain substance adjacent to the mass was reduced to a thin, transparent membrane with irregular linear and patchy deposits of a light gray-brown material which varied from the size of a pinpoint to 7 mm. in diameter. The thinning involved the posterior third of the inferior aspect of both frontal lobes and the adjacent part of the pole of the left temporal lobe. The optic nerves were displaced laterally but were not compressed. The leptomeninges were thin, transparent, and injected. The hypophysis revealed no gross changes.

Upon sectioning the brain, the mass in the region of the infundibulum was found wedged in between the cerebral hemispheres and protruding into the third ventricle. Posteriorly it reached to about 9 mm. from the opening of the sylvian aqueduct, which measured 2 mm. in diameter. The surface of the mass was composed of thin-walled, irregular cavities, the smallest of which was about 8 mm., the largest about 20 mm. in diameter. The cavities were filled with gray-brown, honey-like material. The basal portions of both frontal lobes were covered by a large bilobulated cyst 7.5 cm. in its greatest transverse diameter. The cyst was separated from the brain substance by a light gray-brown membrane about 0.5 mm. in thickness, on the inside of which there was a soft, light yellow-gray, gritty precipitate loosely attached. In the anterior portion of the left half of the cyst there were whitish nodules (5 mm. in diameter), protruding from the wall. The right lateral ventricle was almost completely compressed but did not communicate with the cyst. The left lateral ventricle was slightly dilated.

**Microscopic Examination:** The cyst linings were made up of several layers of small,
flat, elongated cells with oval nuclei which contained many small chromatin granules. The outer layer was formed by a single row of slender and high cylindrical cells, arranged in parallel formation (Fig. 1). Immediately beneath this outer layer, with their axes at right angles to it, were several rows of flattened cells. The innermost of the epithelial layers exhibited several areas in which the epithelial cells formed a loose network. The individual cells were larger and their outlines were angled off so as to form star-shaped structures. Slender protoplasmic bridges connecting the peripheries of adjacent cells were evident. Where the epithelial lining was particularly high, the small elongated cells often showed a tendency to form whorls, containing calcified laminated material in the center. On the inside, the epithelium was covered with irregular, well circumscribed clumps of a partially calcified material, and little toothbud-like outpouchings composed of parallel layers of a material which stained pale purple-pink with hematoxylin-eosin. This homogeneous material seemed to fuse with the uppermost layer of the epithelium. With Heidenhain's iron-hematoxylin stain, the formation of the homogeneous toothbud-like masses was particularly striking. They were found to be derived from epithelial cells that had become greatly swollen through accumulation of a gelatinous intracellular substance (Figs. 1 and 2). Occasionally small granules were discernible. The elongated cells were arranged in band-like lamellae; the nuclear and cell outlines were occasionally evident. In the center were spider-shaped and stellate cells with protoplasmic bridges. At the periphery of these cells, lime salts were deposited in the form of granular material. Often the masses were broken up and separated by accumulation of pus cells and by fibrin and large mononuclear cells containing brownish pigment granules.

In the solid areas, the laminated, homogeneous material was very abundant and showed various stages of calcification. Branched septa composed of the small and elongated cells with a high cylindrical basal layer separated the homogeneous material into irregular islands.

In the region where the cysts bordered on the brain, the epithelial layer rested upon a thin and loose connective-tissue membrane containing many mononuclear cells, lymphocytes, and polymorphonuclear leukocytes. This membrane seemed to be derived from the arachnoid membrane, into which it passed without demarcation.

The structure of the hypophysis was well preserved. The anterior lobe revealed a predominance of the oxyphilic cells. Chromophobe cells were second in frequency, while basophilic cells were scant. At the border of the posterior lobe was a crevice lined by cuboidal cells. A small group of oxyphilic cells were displaced into the adjacent part of the posterior lobe.

Anatomical Diagnosis: Cystic suprasellar adamantinoma invading the third ventricle, with signs of purulent inflammation; surgical osseous defect in the region of the right frontal bone with focal fibrino-purulent external pachymeningitis; fibrous adhesions between the dura mater and both cerebral hemispheres; acute emphysema of the lungs; cloudy swelling and edema of the liver and parenchymatous degeneration of the myocardium and the kidneys.

CASE II: History: C. S., aged forty, Chinese male laborer, entered the Otolaryngology Department of the Cook County Hospital on Aug. 22, 1929, with the diagnosis of chronic otitis media. Because of language difficulties an accurate history could not be obtained. The complaints were apparently difficulty in phonation and pain in the left side of the face and the left eye.

The patient was not acutely ill; the temperature was 99° F., pulse rate 100, and respiratory rate 22. The body showed a scanty distribution of hair, especially in the pubic region. The testicles were markedly reduced in size, while the penis was normal.

Neurological Examination: The pupils were unequal, the right was rigid and larger than the left, which reacted to light sluggishly. There were paralysis of the right internal rectus muscle and slight ptosis of the right eyelid. Nystagmus was not elicited. Vision in the left eye was totally absent; in the right the visual acuity was 0.3. A slight facial paralysis was evident on the left side. The eighth nerve could not be tested because of poor cooperation. The tongue, on protrusion, deviated to the left, and a fine fibrillary tremor was evident. The pharyngeal reflex was present and no paralysis of the laryngeal muscles was demonstrable.
The patellar and achilles reflexes were brisk, the left slightly greater than the right; the abdominal and cremasteric reflexes were both present; pathological reflexes were absent. The power in both upper and lower extremities was equal and good. Sensations were intact throughout.

Laboratory Data: The urine was normal. The blood and spinal Wassermann reactions were negative.

Course: Attacks of profuse epistaxis occurred, followed by bleeding and a foul discharge from the left ear. The patient gradually went down hill, did not yield to supportive measures, and died before blood transfusion could be administered. The clinical diagnosis was basilar syphilitic meningitis.

Necropsy (Dr. R. H. Jaffé): The essential changes were in the brain and skull. Upon removal of the calvarium, the dura mater was seen to be stretched tightly over the cerebral hemispheres. Its surface was smooth and light gray in color. The sagittal sinus contained soft post-mortem blood clots. In removal of the brain, some difficulty was encountered in separating the infundibulum from the base of the skull. In the region of the infundibulum there was a semispherical, light yellow-gray nodule 10 mm. in diameter, which extended from the posterior margin of the chiasm to the middle of the posterior aspect of the infundibulum. The surface of this nodule was smooth. The stalk of the hypophysis was slightly compressed and displaced towards the chiasm. The pons appeared flattened and the basillary arteries were thin-walled and patent. The brain weighed 1370 gm. Over the convexity of the cerebral hemispheres the leptomeninges were separated by an increased amount of fluid. There was a slight diffuse opacity along the meningeal vessels.

The region of the sella was occupied by a soft, friable, pale, pinkish-gray mass, which obscured the diaphragm of the sella and extended into the posterior part of the roof of the left orbit. On the right side the mass reached the ala magna; laterally it was seen extending into the middle fossa, reaching on the left side the opening of the facial canal. Both foramina lacerata were covered by it. Along the facial canal it had grown into the petrous bone, and the anterior and median walls of the left cavum tympani were replaced by it. The cavum was filled with thick, yellowish-green pus, and the membrana tympani had been destroyed. Posteriorly the mass replaced part of the clivus, extending into the region of the jugular veins. From above, the dura mater was seen covering much of the tissue. In places, however, the mass had broken through the dura and was freely exposed after removing the brain.

Fig. 3. Case II: Section of tumor mass showing irregular alveoli surrounded by dense connective tissue at B

S. Spindle and stellate cells in center of mass. C. Cylindrical and cuboidal cells at the periphery. Hemalum-eosin stain. × 75.
Both carotid arteries were firmly embedded in the tissue. Their lumina were narrow but admitted a fine probe. Around the right jugular vein the tissue was destroyed by deep red blood clots.

In the middle cranial fossae the mass was covered by a thin layer of bone. Anteriorly the bone had been completely destroyed. Where the mass bordered on the bone, the latter formed a sharp and indented line. A sagittal section through the middle of the base of the skull revealed a homogeneous, light purplish-gray to grayish-brown tissue which had replaced the sphenoid bone. This tissue filled the sphenoid sinus, the walls of which were laterally expanded. In the posterior part of the sinus there was some soft, whitish medullary tissue. The tuberculum sellae appeared flattened and voluminous, and a light brown tissue was shining through a thin cover of bone. The contour of the anterior lobe of the pituitary gland could be made out, but the posterior lobe seemed to be invaded by the tissue, which grew into the sella from its posterior wall. The roof of the nasopharynx was covered by a thickened mucosa of dirty grayish-green color. The nasopharynx itself was filled by mucoid pus.

FIG. 4. CASE III: MEDIAN SAGITTAL SECTION OF BRAIN SHOWING ENCYSTED TUMOR MASS BULGING INTO THE THIRD VENTRICLE

C. Cyst wall. S. Solid portion of tumor mass.

The intracranial part of the left optic nerve was compressed by the tissue and was thinner than the right.

Microscopic Examination: The tumor of the sphenoid bone was composed of alveoli of various sizes and shapes which showed a tendency to form projections connecting the alveoli with each other. The stroma was of dense fibrillar connective tissue. The alveoli consisted of two types of cells. At the periphery there were cylindrical or cuboidal cells, while in the center spindle-shaped and stellate cells were found. The latter formed a loose network and developed from the former (Fig. 3). Both types of cells were small and possessed dark-staining nuclei. The separation into two zones was especially evident in the larger alveoli, while the small alveoli were made up of cuboidal or polyhedral cells only. The cells revealed no evidence of anaplastic changes or mitosis. There was a marked rarefaction of the bone, the trabeculae of which were separated by connective tissue infiltrated with round cells. It was in this connective tissue that the alveoli of the tumor were found. Some portions of the tumor were necrotic, others were just infiltrated with round cells.

The anterior lobe of the hypophysis was well preserved except for a slight increase of the stroma. The posterior lobe was invaded by the tumor mass which in this location was very densely infiltrated with round cells. The cells of the tumor invaded the intermediary part of the hypophysis, where intracellular blood pigment could be observed. There was also blood pigment in the part of the tumor bordering on the sella.

The whitish mass in the sphenoid sinus proved to be necrotic tumor tissue.

The foregoing structural changes were also present in the nodule attached to the
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The stalk of the hypophysis. It was also densely infiltrated with lymphocytes, larger lymphoid round cells, histiocytes, siderocytes, plasma cells, and polymorphonuclear leukocytes, so that the structure was often obscured. The tumor cells were arranged in the form of an epithelial reticulum. In the center of the nodule an area of loose connective tissue was present.

**Anatomical Diagnosis:** Intrasphenoidal squamous-cell carcinoma of adamantinoma type (hypophyseal duct carcinoma of Erdheim); invasion of the posterior lobe of the hypophysis and local metastasis to the infundibulum; invasion of the right internal jugular vein and the left middle ear; aspiration of blood in both lungs and early bronchopneumonia in the right lower lobe; blood-stained contents of stomach and intestines; chronic edema of the leptomeninges; severe anemia of the internal organs and aberrant pancreatic tissue in the jejunum.

**Case III: History:** G. W., a white man, aged twenty-nine, entered the Research and Educational Hospitals of the University of Illinois on Jan. 14, 1933. The patient had been well until four years before entrance, when, following an automobile accident, he complained of stiffness of the neck and pain on attempting to turn his head to the right. Two years later he injured his jaw; he did not become unconscious. About that time he began to complain of failing vision and entered another hospital, where he was under observation. His vision was decreased, with evidence of central scotomata, and the condition was diagnosed "tobacco amblyopia." Later a right temporal hemianopsia developed with an evident left optic atrophy, but there was no papilledema. The patient left the hospital. After a series of osteopathic treatments his "eyesight improved slightly," but he became very weak and could not move about without assistance. He complained of severe pains in the eyes, head, and back of the neck, and found it difficult to hold his head erect. He was able to lie only on his left side, as other positions caused him pain. His vision became more impaired; he occasionally saw double and two months before his entrance to the Research Hospital had visual hallucinations, seeing pink elephants and alligators. For the last two months he had been drowsy and less active, and he had some slight difficulty in swallowing.

**Examination:** The patient was thin and apathetic; he invariably lay on the left side of his face, his knees drawn up and flexed. He winced when his head was moved, and there was marked suboccipital tenderness. His heart, lungs, abdomen, and genitalia were essentially normal. The left palpebral fissure was slightly wider than the right. The pupils were unequal, the left being larger than the right; neither reacted to light directly or consensually. The external ocular movements were normal. A spontaneous coarse nystagmus to the right was elicited. Ophthalmoscopic examination revealed a bilateral elevation of the disks, of two diopters, with optic atrophy.

The tongue, when protruded, deviated to the left, and the left side of the face appeared to be less mobile than the right. There was a coarse tremor of the upper extremities, especially the right. A slight ataxia was present on the left.

The abdominal reflexes were absent bilaterally; the right cremasteric was active. The knee jerks were exaggerated and a bilateral ankle clonus was present. The muscle power and position sense were intact throughout.

**Laboratory Data:** Spinal and blood Wassermann reactions were negative. The total protein in the spinal fluid was 57 mg. per 100 c.c. The cell count was normal. The urine was normal. The white blood count was 9,800; the red count was 4,700,000.

Roentgenograms taken in antero-posterior and lateral directions showed evidence of destruction involving the posterior boundaries of the sella. Calcification of the pineal body was present, and the antero-posterior view showed no shifting.

**Course:** The history pointed toward a chiasmal lesion, the physical findings, to a cerebellar tumor. A suboccipital exploration was performed (Dr. Eric Oldberg) with negative findings. The patient stood the operation very well. Nothing unusual was seen except that the suboccipital bone was almost paper thin. A chiasmal exploration was postponed for another time.

The patient's temperature rose to 102° F., and the pulse rate to 140. He lapsed into unconsciousness and died on the third postoperative day.

**Necropsy (Dr. G. Milles):** Upon removal of the calvarium, the dura was found tightly
stretched over the cerebral hemispheres. The brain weighed 1250 gms. The meninges were thin and transparent. The convolutions were slightly flattened.

The entire region of the optic chiasm and cerebral peduncles was occupied by a cystic tumor mass which compressed the optic chiasm. The ventral surface of the cystic mass measured 3 cm. in antero-posterior diameter and 2.5 cm. in vertical diameter. On sagittal sectioning of the brain (fixed in a solution of formaldehyde), a tumor mass was seen bulging into the third ventricle, which was markedly dilated (Fig. 4). The interventricular foramina and the lateral ventricles were likewise dilated. The greater part of the tumor consisted of a solid mass containing a fine gritty material and many small cysts filled with yellow-gray material. The walls of the cysts were thickened and their inner surfaces were similarly lined with gritty material, light yellow-gray in color. The mass appeared to arise from the region of the hypophysis, which, however, was intact and distinctly separated from the tumor mass.

FIG. 5. CASE III. SECTION OF TUMOR MASS SHOWING INTERLACING EPITHELIAL COLUMNS AND NUMEROUS CYST FORMATIONS

Concentric whorls seen at C. Van Gieson stain. Low magnification.

Microscopic Examination: The cystic tumor mass was composed of interlacing columns of epithelial cells. These varied considerably in diameter, some were thin and others were several layers thick. The cystic spaces were filled with myxomatous or colloid-like material. The lining of the cysts was formed by a single row of cuboidal cells arranged in parallel formation. Beneath this layer the epithelial cells were indistinct and their nuclei appeared embedded in a sheet of cytoplasm. Occasionally, in the less compact areas, stellate cells were evident. In some areas the epithelial cells formed concentric whorls with a homogeneously staining center and flattened epithelial cells at the periphery (Fig. 5). Calcareous deposits were present in areas about the cysts.

The structure of the hypophysis was well preserved. In the anterior lobe oxyphilic cells predominated.

Anatomical Diagnosis: Intracystic papillary adamantinoma; bronchopneumonia; parenchymatous degeneration of the myocardium; passive congestion of the liver and kidneys.
DISCUSSION AND SUMMARY

In 1892 Onanoff (4) drew attention to the histological similarity of certain tumors of the hypophyseal region to adamantinomas arising from rests of the enamel organ in the jaw. Erdheim (1), in 1904, first suggested that squamous epithelial cells found in this region were the remnants of unobliterated portions of the craniopharyngeal duct, formed from the evagination of the diencephalon which extended downward and came in contact with the buccal evagination of the stomodeum. The tumors composed of these cells were in fact duct tumors (Hypophysenganggeschwülste).

The term ameloblastoma has been suggested instead of adamantinoma by Soy and Churchill (5) and by Frazier and Alpers (6), since it designates the type of cell which comprises the tumor and suggests the resemblance to the fetal structures of the enamel organ. Alpers and Frazier further stated that enamel had never been found in the cases previously described and that the tumor itself possessed no structures that could be said to resemble enamel even remotely.

Dead stratified epithelium variously calcified was described by Duffy (2), who spoke of hydropic phenomena. Farnell (7), Peet (8), and others thought that these masses probably represented atypical or abortive enamel-forming bodies. In a monograph on the histology of tumors of the oral epithelium, Pfüger and Schürmann (9) conclusively described (1931) two cases of adamantinomas in the region of the infundibulum in which definite enamel-forming bodies were present together with embryonic formations of the germ tooth. These tumors, as claimed by the authors, were not of a teratomatous nature and were called Hypophysengangsodontome.

In the first case of adamantinoma reported in this paper, structures were present which were suggestive of enamel formation. These structures consisted of epithelial cell layers which were arranged similarly to the pattern found in the enamel organ. The outer layer was composed of columnar cells having a characteristic palisade formation corresponding to the ameloblasts or gonoblast cells of the enamel organ. Immediately beneath the gonoblasts were several rows of flattened cells, the axes of which lay at right angles, corresponding to the intermediate cell layer of the enamel organ. The innermost cell layer consisted of star-shaped or stellate cells with protoplasmic bridges forming a reticulum corresponding to the pulp or stellate reticulum of the enamel organ. Epithelial pearl formations were present throughout.

Of interest were the toothbud-like projections seen sprouting from the epithelial cells. These cells became greatly swollen and elongated through the accumulation of a gelatinous intracellular substance and revealed small granules in their cytoplasm. These features are suggestive of normal enamel-forming rods. These rods were arranged in the form of lamellae, which later had a marked affinity for calcium salts.

Although there were some structures that resembled enamel-form-
ing cells, they showed considerable deviation from the normal. According to Orbon (10), in the normal formation of the enamel organ there are an outer enamel-forming layer and a stellate reticulum which separates the innermost layer of cylindrical cells constituting the inner enamel epithelium or ameloblast cells. By means of a secretory process, the ameloblastic cells become greatly elongated. Protoplasmic processes or tones develop and later these become calcified, forming the enamel rods at points farthest from their cell of origin.

In this case the elongated rod cells seemed to arise from the masses of epithelial cells and not directly from the ameloblasts. The great majority of adamantinomas develop from the embryonic squamous epithelial rests of the hypophyseal duct either in the infundibulum or beneath the upper surface of the anterior lobe of the hypophysis. Embryologically it is also possible that such tumors may arise between the roof of the pharynx and the sella turcica. In the formation of the hypophysis the buccal evagination loses the connection with the stomodeum by the growth of the sphenoid bone, but cell remnants of the duct may be found even in the adult, in the posterior wall of the nasopharynx, and embryologically, as pointed out by Bailey (11), along the cranio-pharyngeal canal within the sphenoidal bone. From the gross description of the second case here presented, it is evident that the tumor had arisen from within the body of the sphenoid bone, filled the sphenoid sinus, extended into the posterior part of the roof of the left orbit, left middle ear, and right internal jugular vein, and invaded the posterior lobe of the hypophysis, eroding through the dura and giving local metastasis to the infundibulum.

The second case, therefore, reveals distinct evidences of a malignant character. According to Erdheim (1) and Duffy (2), one can distinguish between benign or malignant hypophyseal duct tumors. This malignant character does not manifest itself as much in the microscopic appearance of the cells as in the mode of growth and extension. Thus, the tumor does not differ from the adamantinomas.

In these malignant types of adamantinoma the erosions of the sphenoid bone, the infiltration of the dura, and the base of the brain are usually conspicuous features. According to Duffy's classification the tumor in the third case may be considered an intracystic papillary adamantinoma. The interlacing epithelial columns lined by a single row of cuboidal cells; the central masses composed of stellate cells with a tendency to form whorls; the calcareous deposits and numerous cyst formations—determine the character of this tumor.

Keratohyaline granules were not demonstrable in any of the tumors. The significance of these granules in differentiating the tumors under discussion from epitheliomas of the skin, as emphasized by Erdheim (1) and Jackson (12), can no longer be maintained. Ehlers (13) and Bailey (14) have definitely shown the presence of keratohyaline granules in adamantinomas.

Hair follicles, sebaceous glands, and sweat glands, described by Bostroem (15), Globus (16), Shapiro (17) and others in epidermoid
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cysts simulating hypophyseal duct tumors, were likewise absent in these cases.

Depending upon the stage of formation and differentiation of the oral epithelium, the epithelial cells vary in the formation of the enamel organ. This probably accounts for the variable microscopic appearances of these adamantinomas.

As far as the age of the patients is concerned, the hypophyseal duct adamantinomas are more common in children and adolescents. The youngest patients on record (quoted by Beckman and Kubie, 18) are those of Olivecrona and Lipholm and of Bailey, five and six years of age, respectively. In my first case the patient was only three years of age.

The clinical picture varies with the location of the tumor and with the age of the individual. The symptoms are largely those of intracranial hypertension, disturbance of hypophyseal function, and compression of the neighboring structures. Compression of the optic chiasm is the cause of optic atrophy, defect in the visual fields and visual disturbances. In the three-year-old child there were no marked evidences of pituitary disturbance or pressure on the hypothalamic region except possibly her precocious intelligence. In the second case the scantiness of the pubic hair and the atrophy of the testicles may be mentioned.

The preoperative diagnosis of the tumors of the hypophyseal duct rests largely on the suprasellar calcification which is often detectable on roentgen examination. It is present in the majority of the cases and, when typically developed, is of great diagnostic significance; in fact, it may be considered pathognomonic.

CONCLUSIONS

Tumors of the hypophyseal stalk arise from a definite location, but may show histologic differences. They may present a typical arrangement of the epithelial elements and are usually classified as adamantinomas; some may present atypical enamel formation; others display evidence of malignancy. Cystic degeneration is usually a predominant histologic feature. Clinically the foregoing types can not be differentiated, but a general diagnosis of an extra-hypophyseal tumor can be made.

BIBLIOGRAPHY

11. **BAILEY, PERCIVAL**: Intraeretal Tumors, Springfield, Chas. C. Thomas, 1933.
17. **SHAPIRO, P. F.**: Fetal adenoma of the hypophysis and dermoid cyst of the hypothalamus, Arch. Path. 11: 22, 1931.